RADIOLOGY THROUGH IMAGES

Non-traumatic spleen disorders in children. Assessment by imaging*

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Abstract The spleen is considered a "forgotten organ" by most radiologists and paediatricians despite being affected in many clinical paediatric situations. While it is the organ most often affected in paediatric abdominal trauma, non-traumatic spleen disorders are less well known.

The spleen is well visualised by any imaging technique: ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI); the former is used most often in children. Using imaging techniques to determine the features of splenic anomalies, both congenital and acquired, enables a correct diagnostic approach, avoids unnecessary surgical procedures or biopsies, and helps the clinician to prescribe appropriate treatment.

Our aim was to show the behaviour of the spleen in children using the different imaging techniques: its normal anatomy, the principal anatomical variants and the most common spleen disorder correlating with clinical symptoms, serology and histology.

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PALABRAS CLAVE

Bazo; Ecografía; Tomografía computarizada; Resonancia magnética

El bazo pediátrico no traumático. Valoración por imagen

Resumen Para la mayoría de radiólogos y pediatras, el bazo es el "órgano olvidado", a pesar de estar afectado en múltiples situaciones clínicas de la infancia. Mientras que en el traumaismo abdominal pediátrico es el órgano más implicado, la patología esplénica no traumática es menos conocida.

El bazo se visualiza bien mediante cualquier técnica de imagen: ecografía, tomografía computarizada, resonancia magnética, y de ellas, la primera es la más utilizada en niños. Conocer

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Introduction

The spleen is an intraperitoneal lymphatic organ composed of lymphoid tissue, red pulp and reticuloendothelial cells with two main functions: defensive, through immune response, and blood filtering. It is often affected by generalised infectious, inflammatory, metabolic or neoplastic processes; isolated splenic pathology is rare. Table 1 shows the most common non-traumatic spleen disease in children.

The diagnostic approach to splenic lesions is carried out through a concordant clinical history, a guided serology and the image findings. Ultrasound is the technique of choice in the detection of splenic congenital anomalies, both in the initial study of the pathology and in its follow-up. Computed tomography (CT) and magnetic resonance imaging (MRI) help to characterise the lesions and to plan a conservative surgery of the spleen when required.

Our aim is to show the behaviour of the spleen in the paediatric age group with the different imaging techniques: its normal anatomy, the main anatomical variants and the most common non-traumatic splenic pathology, correlating with clinical, serology or histology.

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Figure 1 Normal spleen of a 5-year-old child. Abdominal ultrasound performed at a hospital admission for acute pyelonephritis. Longitudinal image of the spleen (B), made with a high-frequency linear transducer, showing diffuse reticulonodular pattern of the splenic parenchyma. LK: left kidney.

Normal spleen

The spleen presents a homogeneous parenchyma on ultrasound. High-frequency transducers (12–18 MHz) show the reticulonodular appearance caused by the white and right pulp (Fig. 1). It is common to find intraparenchymal hypoechoic bands forming a zebra-striped spleen (Fig. 2), similar to that seen in CT and MRI during the arterial phase of contrast. However, this ultrasound pattern is not explained by the different vascular flows, but represents the different structural components of the parenchyma.1,2
Congenital malformations

Accessory spleen

This consists of the presence of normal splenic tissue separated from the body of the spleen, produced by failure of the fusion of splenic embryonic buds in the dorsal mesogastrium. Usually less than 3 cm, it can be multiple. The most common location is in the splenic hilum (75%), tail of the pancreas (20%) and splenorenal, splenogastric and splenophrenic ligaments (5%).\(^2\) It has the same behaviour as the main spleen in all imaging techniques. After a splenectomy, the accessory spleen may increase in size and resume the function of the removed spleen.\(^5\) When splenectomy is secondary to haematologic disease (haemolytic anaemia or thrombocytopenic purpura), the presence and location of accessory spleens is essential.

Wandering spleen

The congenital absence, the abnormal development or the laxity of the splenic ligaments allow the spleen to move through the peritoneal cavity with a vascular pedicle which is longer than usual. In up to 64% of cases it presents as an acute abdomen when a splenic infarct secondary to the torsion of the vascular pedicle occurs.\(^7\) By image, the spleen is identified outside the splenic cell, homogeneous if it does not present complications or heterogeneous when the twisted pedicle produces parenchymal infarction. Although splenopexy is the treatment of choice, splenectomy is sometimes required\(^6\) (Fig. 3).

Splenogonadal fusion

This is a rare malformation that occurs in the first weeks of intrauterine life, with persistence of splenic or parenchymal splenic tissue between the spleen and the testicle or left epididymis. The demonstration with any imaging technique of splenic tissue connecting the spleen with the left gonad is definitive.\(^7\)

Figure 2  ‘‘Zebra-striped’’ spleen. A 5-month-old girl who underwent an abdominal ultrasound due to a urine infection. Longitudinal image of the spleen with convex transducer, showing hypoechogenic bands crossing the splenic parenchyma. H: splenic hilum.

It must be remembered that: the ultrasound zebra-stripe pattern is normal in children, it should not be confused with masses and does not require further studies.

In CT, the spleen shows a mottled enhancement in the arterial phase due to the variability of the red pulp flows, and is homogenised in the portal phase. In MRI, the spleen of infants is hypointense in T2 and hyperintense in T1 in relation to the liver, given its predominant composition of red pulp. From 8 months of age, when the lymphoid tissue expands and matures, it acquires the signal pattern similar to that of an adult: moderately hyperintense in T2 sequences and hypointense in T1 with respect to the liver. The pattern of enhancement in MRI is the same as that observed with CT.\(^1\)\(^-\)\(^4\)

Figure 3  Wandering spleen. 3-Year-old boy with diffuse abdominal pain. Upon palpation, a mass can be seen in the left mid-hemiabdomen line. (A) Longitudinal section of B mode ultrasound on left hemiabdomen showing ectopic splenic parenchyma (B) in front of the left psoas muscle (PS). Superficially, serpiginous hypoechogenic structures (white hollow arrow) are distinguished, which in B with linear transducer and colour Doppler mode correspond to vessels forming the swirl sign (white arrow). (C) Abdominal CT confirms the ectopic situation of the spleen (asterisk) with the swirling image of the twisted vascular pedicle (arrow). The homogeneity of the parenchyma is attributed to the fact that, despite the torsion of the pedicle, the infarction is not yet established.
Asplenia/polysplenia

They are part of the spectrum of anomalies known as heterotaxy or cardiosplenic syndromes. Polysplenia presents multiple splenic nodules located in the right or left upper hypochondrium and is associated with abnormalities such as interruption of the vena cava, atresia of the biliary tract or intestinal malrotation. Asplenia is the absence of splenic tissue and is associated with serious cardiac anomalies. Children with asplenia suffer high mortality in the first year of life.\textsuperscript{5}

Splenic cysts

The prevalence of splenic cysts has increased with the widespread use of ultrasound. Between 30\% and 60\% of cases are asymptomatic, and the most common complications are haemorrhage, rupture and superinfection, most likely when the size exceeds 8 cm.\textsuperscript{10,11}

Classically, they are divided into: type I, true or primary, covered by epithelial cells, and type II, pseudocysts or secondary, not covered by epithelium. Other classifications suggest doing so according to their pathogenesis.\textsuperscript{10-12}

Type I, true or primary

- Parasitic (hydatid): the most common in absolute numbers, but not in our environment. Single or multiple, their wall can calcify.
- Non-parasitic: they are more common in the paediatric age group. They can be congenital (epithelial, dermoid, epidermoid) and malformative (lymphatic malformation).

Epithelial cysts are small, less than 4 cm, and can be multiple. By ultrasound they are anechoic, thin-walled, and do not require other imaging studies\textsuperscript{10,11} (Fig. 4).

The epidermoid cysts have a malpighian epithelium (desquamated stratified epithelium) with a cholesterol crystal content in the wall.\textsuperscript{10,11} The ultrasound shows a cystic lesion of bulky size, well delimited, with mobile content and discreetly echogenic by the cholesterol component. The presence of septa or partition walls is common. In computed tomography (CT) they are hypodense with Hounsfield units between 4 and 20, and wall calcifications are observed in up to 10\% of them.\textsuperscript{14} Magnetic resonance imaging (MRI) shows in all the sequences a cystic behaviour with possible uptake of the wall or the septa after contrast\textsuperscript{14,15} (Fig. 5).

The treatment of congenital splenic cysts is recommended in symptomatic cases and in those larger than 4 cm.\textsuperscript{11} The possibilities are wide-ranging, from total or partial laparoscopic splenectomy, to marsupialisation and sclerotherapy, depending on the location and size of the

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**Figure 4** Epithelial cyst 11-year-old girl with recurrent chronic abdominal pain. In ultrasound screening, uncomplicated, unicameral cystic intrasplenic lesion is found. Surgical intervention is considered as the cause of abdominal pain, with the final anatomopathological result of splenic epithelial cyst. PT: pancreas tail.

**Figure 5** Epidermoid cyst. 14-Year-old girl studied with magnetic resonance imaging (MRI) due to spinal anomaly. An injury on the spleen is found as an incidental finding. (A) Coronal section of abdominal MRI SSFSE T2 with presence of 10 × 8 cm lesion, hyperintense, well delimited, with thin incomplete partitions (black arrows) in the interior, which displaces the left kidney (LK) downward. (B) Cross section of abdominal MRI T1 FSE fat sat after gadolinium where the lesion is hypointense, homogeneous, without contrast enhancement, confirming its cystic nature. (C) The ultrasound performed after the MRI study shows the cystic lesion with homogenous echogenic content (white arrows). Surgery was decided because of the large size of the lesion, and laparoscopy was performed with inferior partial splenectomy. The final histological diagnosis is epidermoid cyst. S: spleen.
cyst. More than 25% of the splenic tissue must be saved to preserve its immune function.  

Lymphatic malformations appear as single or multiple cystic lesions filled with lymph. In ultrasound, they are presented as single- or multilocular cystic lesions with septa, anechoic if they are not complicated, of subcapsular location. CT scan and MRI demonstrate its cystic nature and the absence or scarce uptake of its septa after the administration of contrast. They are indistinguishable from the image of the epithelial cysts, and only the histology or the cytology of the intracystic fluid establish the diagnosis.

Type II, pseudocysts or secondary: when they have no epithelial lining or capsule. The most common ones are post-traumatic and post-infectious cysts or after splenic infarcts are much rarer. By imaging they are indistinguishable from type I, so the clinical background is essential for diagnosis.

It must be remembered that: primary cysts are more common in the paediatric age group and their management should be as conservative as possible.

Tumour pathology

Uncommon in the paediatric age group, they appear as incidentalomas. They are divided into benign and malignant.

Benign

**Haemangioma.** It is the most common primary benign tumour of the spleen in children. It can be single or multiple, then associated with cutaneous diffuse haemangiomatosis.

In ultrasound, the behaviour is variable; the most typical is that of a well-defined and hyperechoic lesion, vascularised with the Doppler study. In the CT without contrast, it is iso/hypodense with respect to the surrounding parenchyma, and after administration of intravenous contrast it presents a peripheral enhancement in the arterial phase, with gradual and persistent filling in the venous phase. MRI is the technique of choice to characterise it, since it demonstrates the centripetal and progressively maintained enhancement of the lesion in different phases (Fig. 7). According to its histology, two forms are distinguished: capillary and cavernous. The cavernous cysts are larger, affect a large part of the spleen and can cause severe thrombocytopenia and consumption coagulopathy, giving rise to Kasabach-Merritt syndrome.

**Hamartoma or splenic adenoma.** Splenic hamartomas correspond histologically to an anomalous mixture of splenic components with a predominance of red pulp.

By ultrasound they are solid, hypoechoic with respect to the splenic parenchyma and hypervascular in the Doppler study. In CT they are isodense to the splenic parenchyma, both before and after administration of contrast, and are difficult to detect. Sometimes the only finding of suspicion is an irregularity in the splenic contour (Fig. 8). In MRI they present iso-signal in T1 and hyper signal in T2 with an early, heterogeneous and persistent enhancement over time (Fig. 9).

In symptomatic cases, tumourectomy with partial splenectomy is recommended.

Malignant

Lymphomas and leukaemias. Lymphoma is the most common malignant splenic neoplasm; it can be affected in both Hodgkin’s and non-Hodgkin’s lymphoma. The affected spleen may be normal in size, diffusely enlarged, or

![Figure 6](http://www.elsevier.es/)  
**Figure 6** Lymphatic malformation. 15-Month-old girl with symptoms of crying and abdominal pain. On examination, a mass was palpated in the left hemiabdomen. (A) Ultrasound with a linear transducer on the left hypochondrium showing an intrasplenic multiseptate cystic lesion. (B) Abdominal magnetic resonance, sagittal section SSFSE T2, performed for surgical planning, confirming subcapsular intrasplenic cystic lesion with septa inside of different thickness and presence of capsule (arrows). The histology after splenectomy is lymphatic malformation.
Figure 7  Hamartoma. 11-Year-old boy with abdominal pain of 3 weeks of progression, more accentuated in recent days. (A) Longitudinal section of colour Doppler ultrasound showing a hypoechoic intrasplenic mass (arrowheads), vascularised. (B) Abdominal magnetic resonance imaging (MRI), coronal section SSFSE T2, showing intrasplenic mass (black star) discretely heterogeneous and isointense to normal splenic tissue (white arrows). (C) Cross section, abdominal MRI, dynamic LAVA study in the early phase where the lesion behaves hypervascularly (black star) with respect to the healthy splenic parenchyma (white arrows). (D) In LAVA late phase, the lesion (black star) is homogenised with normal splenic tissue. After laparoscopic splenectomy, the histological result is splenic hamartoma.

Figure 8  Hamartoma. 12-Year-old child with perianal Crohn’s disease. In a flare-up episode of an inflammatory disease, an abdominal ultrasound is performed that reveals a splenic lesion not detected in previous ultrasounds. (A) B-mode ultrasound; longitudinal section of spleen with hypoechoic mass in upper pole (arrow). (B) Cross section of abdominal computed tomography in venous phase; the lesion is isodense to the normal splenic parenchyma and produces only a distortion in the contour of the organ (short arrow). (C) Abdominal magnetic resonance imaging (MRI), coronal section SSFSE T2; the intrasplenic lesion (arrows) is discretely hyperintense with respect to the rest of the spleen. (D) Abdominal MRI, dynamic LAVA coronal section, where the tumour is hypervascular (arrows) in the early phase. The findings suggest splenic hamartoma, and, given the absence of clinical symptoms, it is controlled by ultrasound. The patient remains stable at 5 years of age. LK: left kidney.

have nodular focal lesions that are hypoechoic and typically avascular on ultrasound. Positron emission tomography/computed tomography (PET/CT) is the technique of choice in the staging of lymphoma. Diffuse hypermetabolism of the spleen or focal intrasplenic lesions subcategorises lymphomas as stages (Spleen)\(^9\) (Fig. 10).

Angiosarcoma. It is rare in childhood. It is presented as a heterogeneous mass that in MRI is characteristically hypointense in T1 and T2 with respect to the healthy spleen, with intense contrast enhancement during the arterial phase, persisting in the venous phase and washing in the late phase.\(^{20-22}\)
Haemangioma. 7-Year-old boy with recurring abdominal pain. In abdominal ultrasound screening a splenic lesion is found. (A) B-mode ultrasound longitudinal section of the spleen, with mass in the middle third (arrows), hypoechoic with echogenic centre. (B and C) Abdominal magnetic resonance imaging (MRI), dynamic LAVA cross-section demonstrating the centripetal contrast uptake of the lesion (arrowhead) in the early phase (B) until it is discretely more vascular than the healthy parenchyma in the late phase. (C) After partial splenectomy indicated due to persistence of abdominal pain with no other findings, the histology is splenic haemangioma.

Lymphoblastic lymphoma. 4-Year-old boy with surgical excision of right cervical adenopathy and diagnosis of lymphoblastic lymphoma. Extension study. (A) Longitudinal section of B-mode ultrasound with the presence of multiple intrasplenic hypoechoic diffuse lesions (arrows). (B) PET/CT, coronal fusion section with evidence of hypermetabolism in splenic focal lesions without splenomegaly.

It must be remembered that: benign splenic tumours are rare in the paediatric age group, they are discovered as incidentalomas and their treatment must be conservative.

Infectious pathology

Children with splenic infections have fever, abdominal pain and splenomegaly. The patient’s clinical context, serology and ultrasound findings are sufficient for the diagnosis without requiring other imaging studies. Pyogenic abscesses are rare; the cat scratch disease (Bartonella henselae) should be suspected when splenic hypoechoic nodular lesions coexist with adenopathies in the upper limb and axilla. In immunodeficient children, Candida involvement is the most common, with multiple hypoechoic microabscesses with or without rings in both the spleen and liver. Leishmaniasis, endemic in our environment, shows hypoechoic lesions of variable size, confluent or not, within an important splenomegaly (Fig. 11).
Non-traumatic spleen disorders in children. Assessment by imaging

Figure 11  Infectious pathology. (A) Candidiasis. 3-Year-old girl with pancytopenia and fever. Longitudinal section of colour Doppler ultrasound on the spleen showing splenomegaly with the presence of diffuse hypoechoic millimetric lesions maintaining the vascular architecture. (B) Leishmaniasis. A 5-month-old boy with a fever lasting 7 days and splenomegaly on physical examination. Longitudinal section of B-mode ultrasound of the spleen with the presence of multiple confluent hypoechoic lesions of different sizes, with involvement of even the accessory spleen (asterisk). IgM serology positive for leishmania. LK: left kidney.

Figure 12  Splenic calcifications. 18-Month-old girl with a history of *Bartonella henselae* infection. Longitudinal section of B-mode ultrasound of the spleen with presence of diffuse punctate echogenic calcifications secondary to the previous infectious process. LK: left kidney.

The incidental finding of splenic calcifications usually represents previous infectious processes, mainly due to cat scratch and tuberculosis\(^{14,23,25}\) (Fig. 12).

**Splenic infarction**

Produced by occlusion of the splenic artery or its branches. Haemoglobinopathies are the main cause, and it is less common due to cardiac embolisms, portal hypertension and infiltrative diseases (Gaucher, leukaemia)\(^{2,6}\). The ultrasound shows in the acute phase a peripheral hypoechoic lesion in the shape of a wedge, avascular with Doppler, difficult to differentiate from an abscess or a haematoma. In the chronic phase there is a scar and fibrosis, and it can calcify. The CT shows the extent of the infarction better, since it behaves as a peripheral hypodense zone without enhancement\(^{2,14}\) (Fig. 13).

**Miscellaneous**

*Gaucher disease*. It is a depositional disease caused by deficiency of the enzyme glucocerebrosidase, which causes an accumulation of glucocerebrosides in the reticuloendothelial system. It presents with marked hepatosplenomegaly, either homogeneous or with nodular lesions corresponding to accumulations of Gaucher cells that disappear after enzymatic treatment. Heart attacks and fibrosis are common\(^{14,26}\) (Fig. 13).

*Sickle cell anaemia*. Children with sickle cell anaemia develop acute sequestrum with enlargement of the spleen by vascular engorgement from a young age. Over time,
Figure 14  Homozygous sickle cell disease. 9-Year-old boy, with transfusions since the age of 2. Continuous abdominal pain crisis. (A) Abdominal ultrasound, longitudinal section of spleen with diffuse heterogeneity of the parenchyma due to the presence of Gamna-Gandy bodies together with nodular lesions (asterisks) corresponding to healthy splenic tissue. (B) Cross section of T1 abdominal magnetic resonance within phase, showing nodular lesions (arrows) inside a splenic parenchyma with iron overload. Iron overload also observed in the renal cortex.

Figure 15  Spontaneous splenic rupture. 9-Month-old boy with acute lymphoblastic leukaemia, transplantation from parents, day +13 and sinusoidal obstruction syndrome. (A) Longitudinal section of ultrasound showing distortion of the splenic echostructure and B) abundant echogenic free fluid (FF) due to haemoperitoneum with unstructured splenic inferior pole (arrows). LK: left kidney. The child died after a few hours. The necropsy describes massive splenic rupture.

the spleen becomes self-infarcted and remains small with microbleeds, calcifications and hemosiderin (Gamna-Gandy bodies), together with intrasplenic nodules of functioning parenchyma. If there is transfusion treatment, MRI shows iron overload with a drop in spleen signal for in and out of phase sequences27,28 (Fig. 14).

Spontaneous splenic rupture. It is very rare. It always occurs in a pathological spleen: infectious (infectious mononucleosis, malaria), haematologic (haemolytic anaemia) or neoplastic (leukaemia). Symptoms of acute shock with an unstructured spleen and haemoperitoneum is diagnostic and requires urgent splenectomy19,30 (Fig. 15).

Conclusion

A child’s spleen is affected in a large number of processes. Ultrasound is the technique of choice for its study. Both CT and MRI help the differential diagnosis between splenic lesions of the tumour. The final diagnosis is made by clinical history, serology or histology in certain cases. The treatment of lesions should be as conservative as possible.

Authorship

1. Responsible for the integrity of the paper: CSN.
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10. Approval of the final version: CSN, SPA, RLLS and ECP.

Conflicts of interest

The authors declare that they have no conflicts of interest.

References